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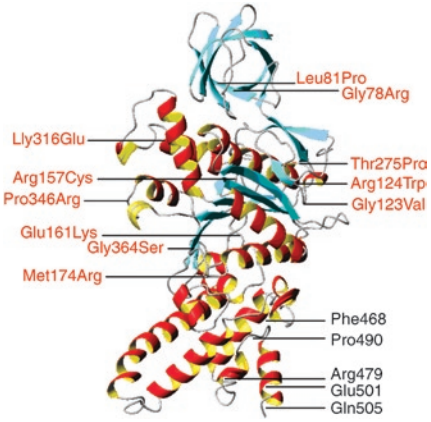
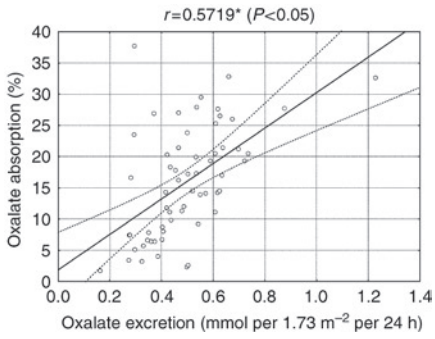
Distal renal tubular acidosis and mutation in the H^+-ATPase



The genetic syndromes that result in distal renal tubular acidosis (RTA) are produced by mutations several genes, including one of the components of the vacuolar H^+-ATPase, the enzyme responsible for H^+ secretion into the urine. The catalytic sector of this enzyme is composed of a large number of subunits, two of which are highly expressed in the intercalated cell of the collecting tubule. Mutations in one such subunit, B1, have been associated with distal RTA. In this issue, Fuster *et al.* report a new mutation in the C terminus of this protein. The authors' elegant biochemical analysis shows that this mutation prevents the assembly of the catalytic sector of the ATPase. They analyzed 12 reported missense mutations in the B1 subunit and found that many of them produced the same phenotype: the failure of assembly, leading to no proton pumping. The assays of proton pumping and pump assembly provide the best possible analysis for the role of these mutations in causing distal RTA. **See page 1151.**

Intestinal oxalate absorption in stone formers

Oxalate, produced by the liver, is present in many foodstuffs. Hence, patients with calcium oxalate nephrolithiasis who excrete large amounts of oxalate in the urine may do so because of overproduction of oxalate by the liver or hyperreabsorption by the intestine. Although the latter can be secondary to intestinal diseases, it is primarily idiopathic. Sikora *et al.* conducted a study in which



^{13}C-oxalate was given to patients with calcium oxalate nephrolithiasis under standard dietary conditions. Calcium oxalate stone formers absorbed larger amounts of oxalate than did controls or patients with primary hyperoxaluria. Oxalate hyperabsorption was seen in one-third of the patients with idiopathic stone disease. Remarkably, patients with idiopathic hyperoxaluria did not hyperabsorb oxalate, suggesting that restricting their oxalate intake is not mandatory except in the case of foods with high oxalate content. **See page 1181.**

Cyclosporine therapy in nephrotic children

Cyclosporine is now a standard treatment for nephrotic syndrome. Ishikura *et al.* studied its efficacy and safety in children with the relapsing variety. A group of patients were given cyclosporine; some received a fixed dose, whereas others received doses adjusted to achieve a trough level of 60–80 ng/ml. In those who received the adjusted dose, the rate of remission was higher and the hazard ratio was lower than in those who received the fixed dose. This finding suggests that managing children with relapsing nephrotic syndrome by maintaining a targeted level of cyclosporine is both effective and safe. **See page 1167.**